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Case Report

Aggressive Angiomyxoma of Vulva: A Rare Entity

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Abstract: Aggressive angiomyxoma (AAM) is a rare locally infiltrative mesenchymal tumour commonly presenting in pelvic and perineal regions in women of reproductive age groups with high risk of local recurrence. So it needs to be differentiated from other mesenchymal tumours of this region. Here, we present the case of a 37 year old woman who had a pedunculated mass over right labia majora with a clinical suspicion of vulvar lipoma.
Keywords: Aggressive Angiomyxoma; Mesenchymal tumour; Vulvar mass.

INTRODUCTION

Aggressive angiomyxoma (AAM) was first described by Steeper & Rosai in 1983 as a distinct myxoid neoplasm in vagina and pelvis (Kaur, A. *et al.*, 2000). The tumour was named aggressive due to its slow and indolent course and high frequency of local recurrence (Brzezinska, B. N. *et al.*, 2018). WHO defines AAM as a tumour of uncertain differentiation (Fletcher C. D. M. *et al.*, 2002). Its prevalence is inconclusive due to its rarity, making management difficult (Wang, Y. F. *et al.*, 2016). Most reports of this disease comes from case reports, with around 350 known cases as of 2012 (Sutton *et al.*, 2012). Considering its aggressive nature and chance of recurrence, complete surgical excision and long term follow up are mandatory.

CASE REPORT

A 37 year old female, para 1, presented with a swelling in right labia majora for 6 months. Local examination revealed a pedunculated skin covered soft and rubbery mass measuring 5 x 5 cm. With a provisional diagnosis of Vulvar lipoma, she underwent local excision of tumour. Cut surface revealed a rubbery, glistening, whitish homogenous appearance (Fig A). Microscopically, there were numerous thick and thin walled blood vessels of varying calibre along with spindled and stellate neoplastic cells in a myxoid to collagenous stroma (Fig B). Alcian blue stain was positive at a pH of 2.5 for the myxoid stroma and Masson's trichrome stained the collagenous stroma (Fig C & D). Postoperative history was uneventful.

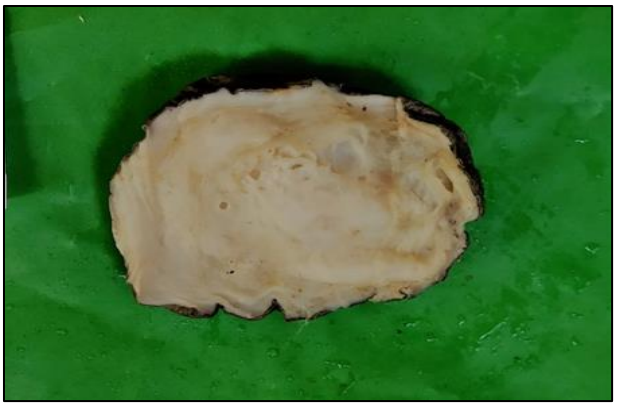

Fig A

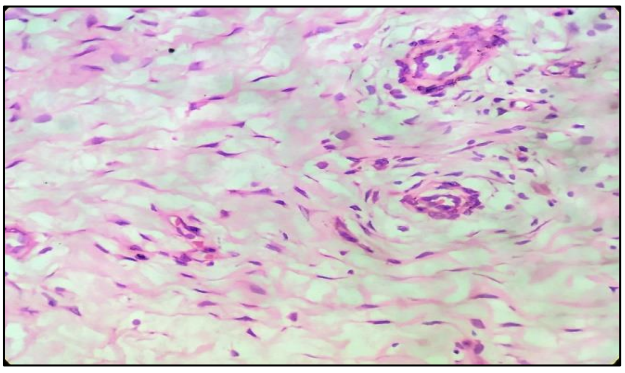

Fig B

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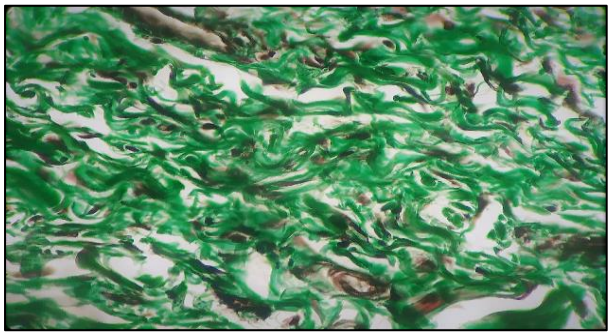

Fig C

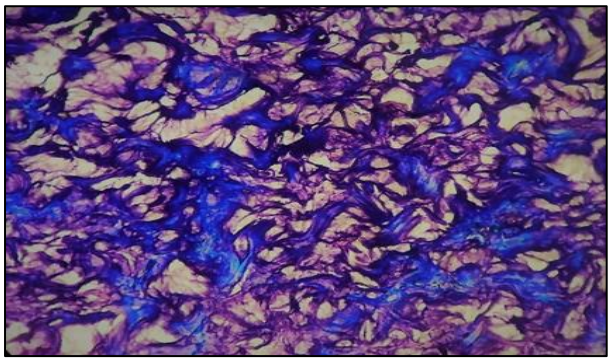

Fig D

Fig A: Gross appearance of AAM. Note the homogenous glistening cut surface. **Fig B:** Photomicrograph showing varying sized blood vessels in a myxoid background (X400) **Fig C:** Massons Trichrome demonstrating collagenous stroma (X400) Fig **D:** Alcian blue positivity of myxoid stroma at pH 2.5 (X400)

DISCUSSION

AAM is a rare but well described tumour commonly occurring in women of reproductive age group with a predilection for pelvic soft tissue, vulva, vagina, rarely the buttocks and inguinal region. A few cases are reported in men (M: F = 1:6) around genital area like perineum, perianal region, scrotum, spermatic cord and inguinal region (Kaur, A. *et al.,* 2000). Clinically it is often diagnosed as Bartholin's cyst, Labial cyst, Lipoma, Gardner's cyst etc. For an accurate and timely diagnosis, it is important to consider this tumour in the differential diagnosis for perineal mass. Misdiagnosis rate is as high as 82% (Bai, H. M. *et al.*, 2013). MRI is the preferred radiological investigation as it shows a distinct "swirling pattern" (Brzezinska, B. N. *et al.,* 2018). Macroscopically, these are soft, rubbery, bulky tumours with glistening homogenous cut surface.

Microscopically, tumour shows thick and thin walled vessels in a myxoid stroma with haphazardly arranged neoplastic stellate and spindle cells. Nuclear atypia and mitosis are rare. Immunohistochemically, it expresses ER, PR, vimentin, CD 34 & SMA and is typically negative for S-100. ER, PR positivity suggests that it is a hormone responsive tumour and is the most characteristic feature (Sutton *et al.,* 2012).

AAM is thought to be arising from specialised mesenchymal cells of pelvic-perineal origin or from a pluripotent perivascular progenitor cell with myofibroblastic and fibroblastic differentiation (Sutton *et al.*, 2012).

Translocation involving Chromosome 12 at 12q 13-15 has been reported in a variety of these mesenchymal tumours. They are thought to affect High Mobility Group A (HMGA2) gene, which is a transcription factor that plays a role in the embryogenesis and not normally detected in adults. This has led to the use of HMGA2 IHC in diagnosing mesenchymal tumours. Recent studies have observed that there is strong nuclear positivity of HMGA2 in 90% of AAM, a weaker positivity in 27% of fibroepithelial polyps and was negative in angiomyofibroblastoma and cellular angiofibroma (Sutton *et al.*, 2012).

Common differential diagnosis of AAM include angiomyofibroblastoma, superficial angiomyxoma, fibroepithelial polyp, myxoid lipomatous tumours and myxoid leiomyoma. The distinctly striking vascular component especially larger vessels with adjacent collagenous and smooth muscle component helps in distinguishing AAM (Chen, H. *et al.,* 2017).

Recurrence rate ranges from 25% to 47% with 85% recurring in initial 5 years (Chen, H. *et al.,* 2017). But distant metastases are rare, except for 3 reported cases of pulmonary metastasis (Brzezinska, B. N. *et al.,* 2018). Hence, complete surgical excision along with hormonal therapy with GnRH agonist / Selective Estrogen Receptor Modulators has been described. Recent literature has shown that radiotherapy is also effective in decreasing tumour size and risk of recurrence (Brzezinska, B. N. *et al.,* 2018).

CONCLUSION

This case report illustrates the diagnostic dilemma while dealing with a vulvar mass. Though AAM is a rare entity, it should always be considered especially in a perimenopausal women. The role of cytochemistry and IHC in AAM is also described.

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